Management of sclerodermal finger ulcers

Ninety-three percent of 59 patients with scleroderma reviewed in this study presented with Raynaud's phenomenon at a mean age of 43 years; 65% of these developed fingertip ulcers within 4 years. Other common findings were sclerodactyly, distal phalangeal resorption, calcinosis cutis, and digital contractures. Medical management of the digital ulcers with systemic and regional vasodilating drugs was unsatisfactory. Sympathectomy, when performed early, temporarily relieved vasospastic pain but did not affect the course of the ulcers. Severe digital pain was the most incapacitating symptom resulting from vasospasm early in the course of the disease and irreversible arterial luminal narrowing later in the course. Conservative fingertip amputations for nonhealing ulcers constituted the management of choice to eradicate the ulcer, to reduce or eliminate the pain, and to return the hand to early useful function. A decision tree for the management of these ulcers is proposed. (J HAND SURG 9A:320-27, 1984.)

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The hands become fixed and the fingers immobile on account of the extreme induration of the skin over the joints. Gradually a diffuse brawny induration develops and the skin becomes firm and hard, and so united to the subcutaneous tissues that it cannot be picked up or pinched. The skin may look natural, but more commonly is glossy, drier than normal and unusually smooth. Bullae and ulcerations have been met with in some instances, and a great deformity of the nails. —Sir William Osler, 1914

This description by Osler of the changes in the hand in scleroderma remains remarkably accurate today. Systemic scleroderma, an uncommon condition, is a disease of connective tissue that may affect the skin, lungs, kidneys, gastrointestinal tract, and heart. Patients with hand involvement present with severe cold intolerance consisting of the triphasic color response of white (vasoconstriction, ischemia), blue (cyanosis, tissue hypoxia), and red (reactive hyperemia), and pain. These vasomotor manifestations associated with vascular spasm in a variety of known diseases are termed "Raynaud's phenomenon"; in contrast, Raynaud's disease represents the idiopathic variety of otherwise similar symptomologies in which no local or systemic causative factors can be found. With progressive involvement of the hands, more than 95% of the patients develop recurrent ulcerations of the fingertips and resorption of the bone in the tufts of terminal phalanges. These ulcerations are superficial moist lesions that are extremely painful and tend to heal very slowly, if at all, leaving depressed scars. In severe involvement of the hands there is progressive flexion contracture of the joints that drastically interferes with the function of the whole hand. No satisfactory management is presently available for these patients with chronic progressive digital ischemia. The conservative treatment employing vasodilators, abstinence from tobacco, and ganglionic blocking agents supplemented frequently by surgical sympathectomy may help some patients only temporarily. Long-term results are discouraging, and fingertip amputations seem to be the only definitive answer for the incapacitating pain these patients experience.

Clinical material

A retrospective chart review was undertaken of all patients diagnosed as having progressive systemic scleroderma and any combination of calcinosis, Raynaud's phenomenon, sclerodactyly, and telangiectases

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Table 1. Incidence of characteristic signs in 59 patients with scleroderma

<table>
<thead>
<tr>
<th>Signs</th>
<th>No. of patients</th>
<th>% of total</th>
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<tbody>
<tr>
<td>Raynaud’s phenomenon</td>
<td>55</td>
<td>93</td>
</tr>
<tr>
<td>Sclerodactyly</td>
<td>50</td>
<td>85</td>
</tr>
<tr>
<td>Esophageal involvement</td>
<td>49</td>
<td>83</td>
</tr>
<tr>
<td>Systemic involvement</td>
<td>47</td>
<td>80</td>
</tr>
<tr>
<td>Finger ulcers</td>
<td>38</td>
<td>64</td>
</tr>
<tr>
<td>Distal phalangeal resorption</td>
<td>35</td>
<td>59</td>
</tr>
<tr>
<td>Calcinosis cuts</td>
<td>28</td>
<td>47</td>
</tr>
<tr>
<td>Hand flexion contractures</td>
<td>17</td>
<td>29</td>
</tr>
<tr>
<td>Other ulcers (toes, trunk)</td>
<td>13</td>
<td>22</td>
</tr>
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</table>

(referred to as the CRST syndrome, or CREST if the esophagus is also involved). The demographic data, symptomatology, and other examinations and treatments were reviewed.

Results

Fifty-nine patients with scleroderma were followed up at the Yale-New Haven Hospital during the 10-year period of 1971 to 1981. Of these patients, 55 (93%) had a history of Raynaud’s phenomenon (Table I). The average age at onset of the phenomenon for these patients was 43 years, with a range of 12 to 69 years (Fig. 1). Of the 59 patients, 38 (64%) developed fingertip ulcers, and these patients exhibited an earlier age at onset of Raynaud’s phenomenon than those patients who never developed finger ulcers. The median age at onset of the digital ulcers was 44 years (range 14 to 70) in women and 52 years (range 38 to 57) in men (Fig. 2). The median period between onset of Raynaud’s phenomenon and the development of finger ulcers was 4.3 years (Table II). Other common symptoms and signs appear in Table I. Diagnostic skin biopsy was performed in 30 patients (51%).

In this group of 59 patients, the female sex predominated 4:1 (48 vs 11 patients). White patients constituted 92% (54 patients) in this series. Although black patients accounted for only 8% of the cases, all of them developed finger ulcers in contrast to only 61% of white patients. Seventy-three percent of all the patients were smokers; however, digital ulcers developed with equal frequency in smokers and nonsmokers. Ten patients with scleroderma were also diagnosed as having one of the following connective tissue diseases: polymyositis, systemic lupus erythematosus, rheumatoid arthritis, and Sjögren’s syndrome.

Fourteen patients died during this 10-year study. Thirteen of these deaths were directly attributed to the patient’s disease, representing an uncorrected mortality of 22% (13/59). Surprisingly, mortality among smokers with scleroderma was only 12% (five patients), while it was 60% (nine patients) among nonsmokers. The age at onset of Raynaud’s phenomenon, development of finger ulcers, or esophageal involvement did not influence the ultimate survival of the patients.

Twenty-eight patients were treated with oral medications during the course of their disease. Among the 13 patients receiving oral reserpine, nine reported no relief while four were afforded subjective relief of their symptoms for periods of less than 2 months. Ten of the patients with fingertip ulcers at the commencement of oral reserpine therapy experienced no resolution of these ulcers even though some patients reported less frequent attacks of pain on exposure to cold. Other drugs that were used frequently were tolazoline and nifedipine. As with reserpine therapy, these drugs led to only a short period of clinical improvement in the circulation of the fingertips.

Twelve patients were treated with intra-arterial injection of reserpine into the brachial artery at 2- to 3-week intervals. Three of these patients experienced
temporary relief of pain for less than 3 months. The intra-arterial reserpine did not heal fingertip ulcers, although most patients did develop a warmer extremity after the injection.

Seven patients were treated with repeated debridement of their hand ulcers, while four were treated with split-thickness skin grafts (Fig. 3). Graft take was complete in three cases; however, these patients experienced no reduction in fingertip pain. The failure of skin graft in one patient was attributed to inadequate debridement. Bone exposure was not present in any of the patients treated with skin grafts. Eight patients underwent cervical sympathectomy; three of these experienced no symptomatic relief, four patients experienced considerable reduction of pain for periods of 1 to 2 years, while one patient had relief of symptoms for about 10 years. Sympathectomies that were deemed successful from the standpoint of resolution of symptoms did not prevent the formation of new finger ulcers, nor did they promote the healing of already established ulcers.

The most successful surgical procedure for the management of fingertip ulcers was amputation of the involved fingertip and primary closure of the stump. This uniformly afforded relief from incapacitating pain, thereby resulting in considerable improvement in hand function.

**Discussion**

**Clinical presentation, incidence, and prognosis.** While a history of Raynaud’s phenomenon was elicited in more than 90% of the patients in this study, the vasospastic pattern was not always present. Several patients described the changes as severe pain upon exposure to cold in conjunction with digital cyanosis or blanching but without reactive hyperemia upon rewarming. Pain persisted for periods of up to 30 minutes after discontinuation of the cold stimulus. This may be explained by the fact that patients with scleroderma have a marked delay in recovery of blood flow after exposure to cold, distinguishing them from patients with Raynaud’s phenomenon attributable to other causes. The morphologic changes in the digital arteries of patients with scleroderma (Fig. 4) and Raynaud’s phenomenon seem to be of prime importance in the development of pain upon exposure to cold especially in advanced stages of the disease. Skin ulcers appear to be indicative of a more rapidly progressing disease. In a study of the morphologic changes in the digital arteries of patients with scleroderma and Raynaud’s phenomenon, Rodnan et al. found these changes in the vessels to consist of severe luminal narrowing as a consequence of severe intimal hyperplasia and adventitial fibrosis while the arterial media was of normal caliber. However, cause-effect relationship of these changes remains unanswered.

All the studies regarding the prognosis in scleroderma are based on a selected sample of patients who had survived to the date of the review. Barnett re-
Fig. 4. A, First digital ulcer in patient with Raynaud’s phenomenon of 3 years’ duration. B, Resolving after 2 months of local care. C, Arteriographic findings of segmental luminal narrowing and complete obstruction. Distal phalangeal tuft resorption is also evident.

Table II. Time periods from onset of Raynaud’s phenomenon to manifestation of digital ulcers and esophageal symptoms

<table>
<thead>
<tr>
<th></th>
<th>With finger ulcers</th>
<th>Without ulcers</th>
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<tr>
<td></td>
<td>Age at onset (yr)</td>
<td>Age range (yr)</td>
</tr>
<tr>
<td>Raynaud’s phenomenon (RP)</td>
<td>39.7</td>
<td>12-69</td>
</tr>
<tr>
<td>Esophageal symptoms</td>
<td>46.6</td>
<td>15-72</td>
</tr>
<tr>
<td>Interval between onset of RP and ulcer development</td>
<td>4.3</td>
<td>0-14</td>
</tr>
<tr>
<td>Interval between onset of RP and esophageal symptoms</td>
<td>6.8</td>
<td>1-37</td>
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</tbody>
</table>

Reported 5- and 10-year survival to be 80% and 55%, respectively. The Mayo Clinic series reported by Tufozelli and Winkelmann and the Hammersmith report by Bennett et al. were similar. However, in the series of patients reported by Rodnan, 7-year survival was less than 35%.

Pathogenesis. In 1924, Matsui reported detailed descriptions of the vascular changes in scleroderma found in complete autopsy studies in five cases. He stressed the constant feature of thickening of the walls and narrowing of the lumina of the small arteries and arterioles in both skin and viscera. Subsequent studies have confirmed that distal arteries and arterioles undergo intimal sclerosis, necrosis, and fibrosis possibly related to ischemia, as observed by arteriographic studies, plethysmography, and the washout method.

The pathogenesis of the observed vascular changes is
Fig. 5. Management of sclerodermal digital ulcers can be separated into four stages: The first stage is from onset of Raynaud’s phenomenon to development of fingertip ulcers. In this stage some show improvement after stellate ganglion block or peripheral nerve blocks and are often relieved of vasospastic pain after sympathectomy and local care. Eventually, new ulcers become resistant to local care. In the second stage, digital vascular damage is overtaking vasospasm as the primary cause of ischemic pain. There may be an indication for cervical or digital sympathectomy. Reserpine therapy every 3 weeks may be an alternative to surgical sympathectomy. Complete absence of vasospastic element brings stage three in which fingertip ulcers are resistant to local care but respond to surgical therapy. In stage four fingertip amputation is inevitable.

unknown but evidence for an immune mechanism is suggested by hypergammaglobulinemia and the frequent occurrence of antinuclear antibody and rheumatoid factor in serum. Circulating immune complexes have been demonstrated by use of the Raji-cell radioimmunoassay, and studies of tissue immunofluorescence reveal immunoglobulin and complement components in the walls of arterioles in the kidneys and lungs of patients dying of scleroderma. Impaired cell-mediated immunity has also been reported. Fibroblasts from patients with scleroderma have been shown to produce collagen faster than do normal skin fibroblasts, and this collagen contains increased amounts of hexosamine compared to normal collagen. An abnormality of monoamine oxidase activity has been suggested in patients with advanced scleroderma.

Medical management

On the whole, the management of scleroderma of the hand has been unsatisfactory. The disease is incurable and progressive, and most treatment regimens can only afford relief of distressing symptoms.

For patients presenting with Raynaud’s phenomenon this should include avoidance of cold and use of warming gloves and mittens in cold weather (Fig. 5); pharmacologic treatment should be used only in those patients who do not respond to cold avoidance. Essential to the local care of ulcers are antibacterial cream and frequent dressing changes. Physical therapy and night splints may maintain muscle strength and delay flexion contractures.

Three groups of drugs have been used in the management of scleroderma cases. These include vasodilators, collagen inhibitors, and immunosuppressants.

Vasodilators. Early in the course of the disease, before the onset of severe arterial stenosis, pharmacologic sympathectomy may be of short-term benefit. The most commonly used agent for this purpose is reserpine administered either intra-arterially or intravenously as a Bier block. Porter et al. demonstrated that intra-arterial injection of reserpine results in depletion of arterial wall catecholamines for 14 to 21 days. Although Nilsen and Jayson reported that repeated injections of reserpine relieved the pain and contributed to the healing of indolent ulcers of the fingers, this is at variance with our experience and with the experience of others. We have found that although intra-arterial reserpine may result in reduction of pain and improvement of circulation, evidenced by persistent erythema and elevated skin temperatures (Fig. 6), the course of the ulcers has not been influenced.

Other peripheral vasodilating agents that have been used include nitroglycerin paste applied to the forearm before exposure to cold, oral phenoxybenzamine, nifedipine, guanethidine, and alpha-methyldopa. Prostaglandin E1, a vasodilator and potent inhibitor of plate-
**Table III. Criteria for cervical or digital sympathectomy**

<table>
<thead>
<tr>
<th>Improvement in color, temperature, perfusion, number of vasospastic attacks, chronic digital pain, and resolution of ulcers in the hand following:</th>
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<tbody>
<tr>
<td>1. Slow intra-arterial (brachial artery) administration of 1.25 mg of reserpine</td>
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<tr>
<td>2. Intravenous administration of 1.25 mg of reserpine by use of Bier block technique</td>
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<tr>
<td>3. Stellate ganglion block with bupivacaine</td>
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<tr>
<td>4. Combined radial, ulnar, and median nerve blocks at the level of the wrist by use of bupivacaine</td>
</tr>
<tr>
<td>5. Common and proper digital nerve blocks in web spaces adjacent to the involved digit by use of bupivacaine</td>
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</table>

LET aggregation, administered intravenously for 72 hours has been reported to result in short-term relief of pain, healing of fingertip ulcers, and increased perfusion of fingers for up to 6 weeks.20

**Collagen inhibitors.** The use of these drugs is aimed at the inhibition of connective tissue biosynthesis. While D-penicillamine, an agent known to interfere with cross-linking of collagen fibrils, has been used with variable success,20 colchicine, which inhibits collagen secretion from cultured fibroblasts in vitro, has not demonstrated consistent clinical efficacy.20

**Immunosuppressants.** These include corticosteroids, cyclophosphamide, chlorambucil, melphalan, and azathioprine. Unfortunately, results have been inconclusive at best,21,22 and the relationship of autoimmunity to scleroderma requires further clarification.

While treatment regimens focusing upon alterations of collagen synthesis and modulation of humeral and cellular immunity have a sound theoretic basis, they remain largely empirical and appear to result in only short-term symptomatic relief.

**Surgical management**

The aspects of scleroderma of the hand that are amenable to surgical management are vasospastic pain, refractory digital ulcerations, and flexion contractures (Fig. 5). In terms of priority, the successful treatment of hand pain will enable the patient to live a more comfortable life and be better able to cope with this progressive and incurable disease.

**Vasospastic pain.** Sympathectomy is the most frequently employed operation in the management of the pain. Early reports of its use in scleroderma were published by Adson et al.,23 who successfully treated 16 patients, and by Leriche et al.,24 who reported significant improvement in nine of 13 patients. More recently, Barnett and Coventry25 reported sympathec-

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**Fig. 7.** A, Painful nail bed ulcer in patient with progressive systemic sclerosis. B, Three months later the ulcer is larger despite local care and intra-arterial injections of reserpine every 3 weeks. Amputation at proximal interphalangeal level followed, with complete relief of pain.

Sympathectomy to be of value with respect to relief of ischemic symptoms in 12 of 20 patients. Other studies also report short-term improvement from sympathectomy.26 The reason for the complete failure of cervical sympathectomy in some patients may be related to the fact that the brachial plexus does not receive its communicating rami exclusively from the cervicothoracic sympathetic trunk; additional sympathetic fibers may reach the brachial plexus through the sinuvertebral nerve, the arnold plexus, and the nerve of Kuntz. Because of these variations in proximal sympathetic contributions, Flatley27 reported a technique of digital sympathectomy for use in distal circulatory problems. He reported substantial relief of pain with a persistent temperature rise of 2°F in the finger of one patient with scleroderma who had undergone digital sympathectomy, but he observed no healing of the ulcerated fingertips. Wilgis,28 however, reported complete healing of digital ulcers in 8 of 9 patients after digital sympathectomy. Improvement subsequent to a stellate ganglion block (chemical sympa-
thectomy) may be evidence for possible benefit from a surgical cervical sympathectomy (Table III), whereas a positive response to peripheral blockade with bupivacaine may be an indication for digital sympathectomy.38

In late scleroderma the digital arterial perfusion pressure has been found to be about 25% of normal39 and no difference could be demonstrated between sympathetic- and nonsympatheticized fingers. These observations support the conclusion by Lewis and Landis40 that the prime factors in the ischemic changes of late scleroderma are due to local faults of the blood vessels and not to the sympathetic nervous system, in contrast to early scleroderma where the order of importance is reversed.

**Digital ulcers.** When noninvasive evaluation of the digital ischemia by Doppler examination, pulse volume recordings with cold stress, sympathetic blockade,38 and radionuclide imaging of the hand41 are inconclusive, arteriography may be performed to determine the status of the digital vessels (Fig. 5).

The important interval management of digital ulcers are surgical debridement of nonviable tissue and topical application of penetrating antibacterial agents such as silver sulfadiazine for the prevention of local infection. Skin grafting has been used in the management of joint contractures, scalp defects, and trunk wounds of scleroderma patients. Fries et al.42 demonstrated that when clinically normal skin was placed on a sclerodematous bed, the graft became sclerodermatous; and sclerodermatous skin placed in a normal bed remained diseased. Successful skin grafting does not seem to reduce or eliminate pain in most patients. It was the unrelenting pain of this nature that led to narcotic dependence in 40% of the patients in our series with digital ulcers.

At present, the only treatment that provides the potential for both healing the wound and relieving the pain in patients with persistent digital ulcers is a planned partial amputation of the digits (Fig. 7). An arteriogram may help the surgeon select the appropriate level of amputation and avoid recurrence of the ulcers in the early postoperative period. The amputation site usually heals surprisingly well, an observation also reported by others.43 In our series, no problems in wound healing were observed in the stumps of 25 partial digital amputations.

**Flexion contractures.** Reconstructive surgery is effective for repairs of deformities, severe contractures, and stiffness that seriously limit the use of the hand in these patients. The skin incision and arthrodeses usually heal without complication. It has been suggested by Lipscomb et al44 that the proximal interphalangeal joints should be fused in 55° to 60° of flexion rather than the conventional 30° to 45° so that the thumb will oppose those fingers with the limited motion. The use of silicone rubber prostheses in the metacarpophalangeal joints and osteotomies of the deformed fingers have also been reported by Entin and Wilkinson.4

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Management of scleroderma finger ulcers

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